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ONLINE SUPPLEMENTARY TABLES:

Table S1: Definitions for comparator groups

Comparator Group	Applied definition in DCVAS database
Giant Cell Arteritis Comparator Group	<ul style="list-style-type: none">• Presenting clinical scenario of New-onset headache, sudden visual loss, ischaemic jaw or tongue pain AND• Age > 40 years at onset of symptoms
Takayasu's Arteritis Comparator Group	<ul style="list-style-type: none">• Presenting clinical scenario of Aortic aneurysm, new-onset hypertension associated with other systemic features, Stroke, Limb claudication, chronic headache AND• Age ≤ 50 years at onset of symptoms
ANCA-Associated Vasculitis + PAN Comparator Group	<ul style="list-style-type: none">• Presenting clinical scenario of acute and progressive renal impairment and/or failure, Haemoptysis/pulmonary haemorrhage, Acute respiratory distress, exacerbation of asthma or unexplained pulmonary fibrosis, new-onset hypertension associated with other systemic features, Peripheral blood eosinophilia, peripheral neuropathy, inflammatory polyarthritis, acute or chronic abdominal pain
IgA Vasculitis Comparator Group	<ul style="list-style-type: none">• Presenting clinical scenario of acute or progressive renal impairment and/or failure, Rash/skin abnormalities (including nodules and ulcers), Acute or chronic abdominal pain

Table S2: Definitions for each individual criterion in the ACR classification criteria sets

ACR-Criterion	Applied definition in DCVAS database (any of the definitions per criteria, unless specified otherwise)
GIANT CELL ARTERITIS	
1. Age \geq 50 years	<ul style="list-style-type: none"> Age \geq 50 years at time of diagnosis
2. New onset headache	<ul style="list-style-type: none"> Presenting clinical scenario of new onset headache Clinical feature of headache regardless of specific location
3. Temporal artery abnormalities	<ul style="list-style-type: none"> Diminished pulse, tenderness over the temporal artery or cord-like-sign over left or right temporal artery
4. ESR \geq 50 mm/h	<ul style="list-style-type: none"> Erythrocyte sedimentation rate \geq 50 mm/h
5. Abnormal biopsy	<ul style="list-style-type: none"> Biopsy of the temporal artery, aorta or other artery with Findings consistent or diagnostic of vasculitis
TAKAYASU'S ARTERITIS	
1. Age \leq 40	<ul style="list-style-type: none"> Age \leq 40 years at time of diagnosis
2. Claudication	<ul style="list-style-type: none"> Presenting clinical scenario of limb claudication OR Leg or Arm claudication
3. Decreased brachial artery pulse	<ul style="list-style-type: none"> Either diminished or absent pulse of brachial artery on vascular examination
4. Brachial artery blood pressure difference $>$ 10mmHg	<ul style="list-style-type: none"> Brachial artery blood pressure difference $>$ 10mmHg
5. Bruit over subclavian artery or aorta	<ul style="list-style-type: none"> Bruit over subclavian artery or aorta on vascular examination
6. Arteriogram abnormalities	<ul style="list-style-type: none"> Vessel narrowing or vessel occlusion in any large artery detected on catheter-based angiogram, CT(-angiogram), MR(-angiogram), fluorescein-angiogram or ultrasound In the absence of calcification of the vessel involved
GRANULOMATOSIS WITH POLYANGIITIS (WEGENER'S)	
1. Nasal / oral inflammation	<ul style="list-style-type: none"> Bloody nasal discharge Non-blood stained nasal discharge Mouth ulcers
2. Abnormal chest x-ray	<ul style="list-style-type: none"> Nodules or consolidation, cavities, infiltration, inflammation or pleural thickening on CT, MRI or x-ray of the chest
3. Urinary sediment	<ul style="list-style-type: none"> Blood on urine dipstick or Red cell casts in urine on urine-microscopy
4. Granulomatous inflammation on biopsy	<ul style="list-style-type: none"> Biopsy regardless of site showing granuloma or extravascular granulomatous inflammation
EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (CHURG-STRAUSS)	
1. Asthma	<ul style="list-style-type: none"> Positive history of asthma
2. Blood eosinophilia $>$ 10%	<ul style="list-style-type: none"> Blood eosinophilia $>$ $1.5 \times 10^9/l$
3. Neuropathy	<ul style="list-style-type: none"> Presenting clinical scenario of peripheral neuropathy (either sensory or motor) or Mononeuritis multiplex or Motor neuropathy (not due to radiculopathy) or Sensory neuropathy (not due to radiculopathy) Confirmation by EMG/NCS was not required
4. Non-fixed pulmonary infiltrates	<ul style="list-style-type: none"> Infiltration, consolidation or inflammation on CT, MRI or x-ray of the chest
5. Paranasal sinus abnormalities	<ul style="list-style-type: none"> Non-blood stained nasal discharge or Sino nasal congestion or blockage or Inflammation, effusion or polyps in the paranasal sinuses on CT, MRI or x-ray.
6. Extravascular eosinophilic infiltration	<ul style="list-style-type: none"> Biopsy regardless of site showing extravascular eosinophil-predominant inflammation
POLYARTERITIS NODOSA	
1. Weight loss \geq 4kg	<ul style="list-style-type: none"> Weight loss \geq 5kg
2. Livedo reticularis	<ul style="list-style-type: none"> Livedo reticularis on examination

ACR-Criterion	Applied definition in DCVAS database (any of the definitions per criteria, unless specified otherwise)
3. Testicular pain or tenderness	<ul style="list-style-type: none"> • Testicular pain or • Testicular tenderness on examination
4. Myalgia, weakness or leg tenderness	<ul style="list-style-type: none"> • Myalgia (muscle pain) or muscle cramps or • Muscle weakness on examination or • Muscle tenderness on examination • In the absence of a presenting clinical scenario of inflammatory shoulder and/or hip girdle symptoms (polymyalgia-like symptoms)
5. Mono- or polyneuropathy	<ul style="list-style-type: none"> • Presenting clinical scenario of peripheral neuropathy (either sensory or motor) or • Mononeuritis multiplex or • Motor neuropathy (not due to radiculopathy) or • Sensory neuropathy (not due to radiculopathy) • Confirmation by EMG/NCS was not required
6. Diastolic blood pressure > 90 mmHg	<ul style="list-style-type: none"> • Systolic Blood Pressure > 140 mmHg
7. Elevated blood urea nitrogen or creatinine (>1.5 mg/dl)	<ul style="list-style-type: none"> • Creatinine > 130 µmol/l
8. Presence of HBV-surface antigen or antibody in serum	<ul style="list-style-type: none"> • Hepatitis B (evidence of acute or ongoing chronic infection)
9. Arteriographic abnormality	<ul style="list-style-type: none"> • Vessel occlusion, aneurysm, beading or micro aneurysms of the mesenteric arteries demonstrated by catheter-based dye angiogram, CT(-angiogram) or MR(-angiogram)
10. Biopsy of small or medium-sized artery containing PMN	<ul style="list-style-type: none"> • Biopsy of a small or medium sized artery with predominantly neutrophilic vasculitis or predominantly mononuclear leukocytes in vasculitis
IGA VASCULITIS (HENOCH-SCHÖNLEIN)	
1. Palpable purpura not related to thrombocytopenia	<ul style="list-style-type: none"> • Palpable purpura on examination without presence of thrombocytopenia (<100x10⁹/l)
2. Age ≤ 20 years at disease onset	<ul style="list-style-type: none"> • Age ≤ 20 years at onset of symptoms
3. Bowel angina	<ul style="list-style-type: none"> • Abdominal pain (any) • Postprandial abdominal pain / ischaemic abdominal pain • Peritonism on examination • Mesenteric ischaemia
4. Wall granulocytes on biopsy	<ul style="list-style-type: none"> • Biopsy presenting • Predominantly mononuclear leukocytes in vasculitis or • Necrotizing or leukocytoclastic arteriolitis or • Necrotizing or leukocytoclastic venulitis

Table S3: Overview of clinical conditions in the comparator patients

Comparator conditions	GIANT CELL ARTERITIS n=81	TAKAYASU'S ARTERITIS n=24	ANCA-ASSOCIATED VASCULITIDES +PAN n=267	IgA-VASCULITIS n=228
Dermatologic	0	1	5	13
Non-ischæmic skin-lesions				3
Other dermatologic condition*		1	5	10
Endocrinologic/metabolic	1	0	1	2
Thyroid disease	1			
Pituitary macroadenoma			1	1
Other endocrinologic conditions			0	1
Gastrointestinal	0	0	9	10
Cholecystitis			1	1
Eosinophilic esophagitis			2	
Other gastroenterologic conditions			4	10
Genitourinary	0	0	7	4
Nephrotic / nephretic syndrome			5	4
Interstitial tubular nephritis			1	1
Acute kidney injury			1	1
Focal progressive IgA-Nephritis			1	1
Haematologic	2	1	8	10
Systemic amyloidosis	1		1	1
Coagulopathy / Thrombosis		1		1
Hypereosinophilic syndrome			3	2
Myeloma / Paraproteinaemia			3	3
Monoclonal cryoglobulinaemia			1	
Other hematologic	1			3
Infectious diseases	3	1	23	26
Bacterial endocarditis	1		8	7
Pneumonia (bacterial / viral)		1	2	1
Upper respiratory infection				1
Urinary tract infection			3	3
Eosinophilic pneumonia			1	
Streptococcal Group A infection			2	
Other infections	2		7	14
Malignancy	2	0	4	5
Solid malignancy	2			1
Haematologic malignancy			4	4
Neurologic	26	5	7	3
Migraine / headache syndrome	21	1	2	1
Neuropathy not due to vasculitis	1		2	
Stroke not due to vasculitis	1	1		1
Multiple sclerosis		1	1	1
Other neurologic conditions**	5	2	2	
Ophthalmologic	10	0	1	0
Vision loss not due to vasculitis	8			
Other ophthalmologic conditions	2		1	1
Other	4	0	7	12
Respiratory	1	0	20	9
Asthma			6	4
Allergic eosinophilic pneumonia			2	
COPD			1	
Interstitial lung disease			7	3
Other respiratory conditions***	1		4	2
Rheumatologic	25	10	158	105
SLE	3	6	40	34
Rheumatoid arthritis	1	0	27	10
Sarcoidosis	0	0	14	8
Dermatomyositis / Polymyositis	2	0	11	13
Sjogren's syndrome	4	0	5	2
Periodic fever syndrome	0	1	3	5
Antiphospholipid-syndrome	0	1	1	1
Adult onset Still syndrome	1		4	4
Anti-synthetase syndrome			3	2
Polymyalgia rheumatic	2		5	1
IgG4-related disease		1	6	2
Other rheumatologic ****	7		12	10
Other CTD*****	0	0	16	11

Table S3: Overview of clinical conditions in the comparator patients [continued]

Comparator conditions		GIANT CELL ARTERITIS n=81	TAKAYASU'S ARTERITIS n=24	ANCA-ASSOCIATED VASCULITIDES n=267	IGA-VASCULITIS n=228
Other inflammatory arthritis		5	1	14	3
Vascular		5	6	9	13
Artherosclerosis		3	3	3	1
Thrombangiitis obliterans					1
Other vascular conditions		2	3	6	11
Toxic		0	0	5	11
Propylthiouracil induced vasculitis				3	
Other drug-induced				2	
*	including bullous pemphigoid and unspecific dermatitis				
**	including Bell's palsy, Moyamoya and subarachnoidal haemorrhage, lumbosacral radiculopathy				
***	including cryptogenic organizing pneumonia, granulomatous disease				
****	including relapsing polychondritis, eosinophilic fasciitis, fibromyalgia				
*****	including overlaps of systemic sclerosis with SLE and poly-/dermatomyositis				

Table S5: Differences between patients who were classified by the 1990 ACR criteria in accordance with physician's submitted diagnosis or not with respect to fulfilment of individual ACR criteria

ACR-Criterion	Correctly classified by the ACR criteria		<i>p-value</i>
	YES	NO [#]	
GIANT CELL ARTERITIS (n=345)	280 (67.1)	65 (32.9)	
1 - Age > 50 years	278 (99.3)	60 (92.3)	<0.001
2 - Headache	249 (88.9)	17 (26.2)	<0.001
3 - Temporal artery abnormalities	130 (46.4)	2 (3.1)	<0.001
4 - ESR ≥ 50	217 (77.5)	37 (56.9)	0.001
5 - Positive temporal artery biopsy	204 (72.9)	5 (7.7)	<0.001
TAKAYASU'S ARTERITIS (n=53)	39 (73.6)	14 (26.4)	
1 - Age ≤ 40 years	28 (71.8)	8 (57.1)	0.314
2 - Claudication	26 (56.6)	4 (28.6)	0.014
3 - Reduced brachial pulse	17 (43.6)	1 (7.1)	0.014
4 - Difference in brachial artery blood pressure	16 (41.0)	0	0.004
5 - Bruits (aorta or subclavian artery)	27 (69.2)	2 (14.3)	<0.001
6 - Arteriogram abnormalities	34 (87.2)	8 (50.0)	0.004
GRANULOMATOSIS WITH POLYANGIITIS (n=275)	181 (65.8)	94 (34.2)	
1 - Oral / nasal Inflammation	138 (76.2)	27 (28.7)	<0.001
2 - Abnormal chest imaging	117 (64.6)	17 (18.1)	<0.001
3 - Urinary sediment	133 (73.5)	28 (30.8)	<0.001
4 - Positive biopsy	51 (28.2)	6 (6.4)	<0.001
EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (n=97)	45 (57.0)	34 (43.0)	
1 - Asthma	34 (75.6)	18 (52.9)	0.036
2 - Eosinophilia	40 (89)	25 (73.53)	0.077
3 - Neuropathy	39 (86.7)	20 (58.8)	0.005
4 - Abnormal chest imaging	28 (62.2)	4 (11.7)	<0.001
5 - Paranasal sinus abnormalities	37 (82.2)	18 (50)	0.002
6 - Positive biopsy	14 (31.1)	1 (2.9)	0.002
POLYARTERITIS NODOSA (n=32)	13 (40.6)	19 (59.4)	
1 - Weight loss > 4 kilograms	7 (53.9)	4 (21.1)	0.055
2 - Livedo reticularis	3 (23.1)	1 (5.3)	0.135
3 - Testicular tenderness / pain	4 (30.8)	0	0.01
4 - Myalgia, weakness, leg tenderness	10 (76.9)	5 (26.3)	0.005
5 - Mono- / polyneuropathy	8 (61.5)	4 (21.1)	0.02
6 - Diastolic blood pressure > 90mmHg	4 (30.8)	3 (15.8)	0.314
7 - Elevated BUN or Creatinine	3 (23.1)	1 (5.3)	0.135
8 - Presence of HBV-infection	4 (30.8)	1 (5.3)	0.051
9 - Arteriographic abnormalities	1 (7.7)	1 (5.7)	0.78
10 - Biopsy of vasculitis with PMN	1 (7.7)	2 (10.5)	0.787

Table S4: Number of patients recruited per centre and centre-specific sensitivity and specificity of the 1990 ACR classification criteria for each form of PSV. 24 of 85 centres with at least 15 PSV patients with primary systemic vasculitis are displayed.

Centre	Country	Speciality	PSV patients																					Other PSV	Comparators	
			All		GCA		TAK		GPA		MPA*		EGPA		PAN		IgAV			n	n					
			n	n	Sens	Spec	n	Sens	Spec	n	Sens	Spec	n	Sens	Spec	n	Sens	Spec	n			Sens	Spec			
1	SI	Rhe	89	44	91	97			97	9	78	90	1	100	95	4	100	100	2	0	96	29	76	93	13	21
2	ENG	Rhe/Neph	61	35	91	93	2	50	100	18	50	100	2	50	97	1	100	100			94	3	100	100	1	27
3	ENG	Rhe	49	31	81	96	2	0	100	11	73	94			78	3	67	98	1	100	79	1	100	98	10	59
4	GER	Rhe	41	22	50	87	4	50	95	13	62	97			96			100	2	100	100			100	4	35
5	USA	Rhe	37	10	90	85	4	50	100	10	40	93	4	25	85	7	0	100	1	0	78	1	0	97		7
6	RU	Neph	34	2	100	100	2	50	100	16	56	100	1	0	100	10	40	100			97	3	0	100		35
7	GER	Rhe	33	2	50	100	2	50	100	24	79	100	3	0	91	1	0	100			83	1	0	97	2	1
8	ENG	Rhe	32	26	85	75			100	4	50	93	1	100	97			100			94	1	0	97	2	
9	ENG	Rhe/Oph	31	22	100	100			100	5	40	90	4	25	90			100			79			100	3	11
10	DK	Rhe	31	8	63	100	5	80	96	17	29	100			100			100			100	1	100	94	1	3
11	CH	Rhe	26	22	59	100			97	2	50	100	2	0	93			100			87			97	5	24
12	CAN	Rhe	26	8	88	95			100	8	63	74			89	7	57	100			89	3	100	92	1	20
13	CAN	Rhe	26	3	100	100			100	11	55	65	6	33	86	1	100	100	4	25	79	1	0	96	2	4
14	ENG	Rhe	25	15	87	79			100	5	80	96	2	50	96	2	0	100			90	1	100	100	4	16
15	CAN	Rhe	25	5	100	81	2	100	90	1	100	80	4	50	78	12	75	100	1	100	70			90	6	9
16	PL	Int Med	18	1	100	86			100	8	88	60	3	33	75	5	100	100			65	1	100	91	5	6
17	TK	Rhe	18	3	100	93	7	100	100	1	0	94	2	50	100			100	1	0	94	4	100	100		2
18	BEL	Int Med	17	10	80	100			100	5	100	92	1	0	94			100	1	0	94			100		1
19	JP	Neph	16	1	100	93			100	2	50	71	12	50	100			100	1	0	27			100		8
20	IT	Rhe	16	4	50	93	1	100	100	4	50	100	1	0	89	3s	100	100	3	33	100			95	3	
21	CN	Rhe	15	1	0	100	5	40	88	4	100	88	2	0	84	1	0	100	2	50	84			95	6	14
22	SCO	Rhe	15	2	0	94			100	8	88	82	1	100	83	3	33	100	1	0	83			100	4	13
23	CN	Neph	15			94			100	5	100	69	10	60	50			100			22			94	3	1
24	PT	Neuro	15	10	100	100	2	100	100	1	100	100	1	0	93			100	1	100	93			100		

PSV: primary systemic vasculitis; Sens: Sensitivity; Spec: Specificity; Rhe: Rheumatology; Neph: Nephrology; Ophth: Ophthalmology; Int Med: Internal medicine; Neuro: Neurology; GCA: giant cell arteritis; TAK: Takayasu's arteritis; PAN: polyarteritis nodosa; MPA: microscopic polyangiitis; GPA: granulomatosis with polyangiitis; EGPA: eosinophilic granulomatosis with polyangiitis; IgAV: IgA-vasculitis. *Sensitivities and specificities are shown for PAN criteria applied to MPA patients (details given in the Methods)

Table S5: Differences between patients who were classified by the 1990 ACR criteria in accordance with physician's submitted diagnosis or not with respect to fulfilment of individual ACR criteria [continued]

MICROSCOPIC POLYANGIITIS (n=94)	27 (28.7)	67 (71.3)	
1 – Weight loss > 4 kilograms	15 (55.6)	10 (14.9)	<0.001
2 - Livedo reticularis	4 (14.8)	2 (3.0)	0.034
3 - Testicular tenderness / pain	0	0	-
4 - Myalgia, weakness, leg tenderness	15 (55.7)	10 (14.9)	<0.001
5 - Mono- / Polyneuropathy	15 (55.7)	12 (17.9)	0.001
6 - Diastolic Blood Pressure > 90 mmHg	19 (70.4)	24 (35.8)	0.002
7 - Elevated BUN or Creatinine	19 (70.4)	41 (61.2)	0.402
8 - Presence of HBV-infection	0	0	-
9 - Arteriographic abnormalities	0	0	-
10 - Biopsy showing vasculitis with PMN	2 (7.4)	1 (1.5)	0.14
IGA-VASCULITIS (HENOCH-SCHÖNLEIN) (n=66)	48 (72.7)	18 (27.3)	
1 - Palpable purpura	47 (97.9)	15 (83.3)	0.027
2 - Age ≤ 20 years at onset	2 (4.2)	0	0.379
3 - Bowel angina	27 (56.3)	3 (16.7)	0.004
4 - Wall granulocytes on biopsy	31 (64.6)	0	<0.001
Definitions applied for each of the criteria are shown in Supplementary Table S2 . Differences between criteria fulfilment were analysed using Pearson- χ^2 -test. All p-values are two-tailed and were considered significant if < 0.05.			
#not correctly classified by 1990 ACR criteria means that either they did not meet the criteria or were classified with a diagnosis not concordant with the one submitted by the investigator.			
ACR: American College of Rheumatology, BUN: blood urea nitrogen, ESR: erythrocyte sedimentation rate, HBV: hepatitis B virus, PMN: polymorphonuclear leukocytes			